PRIMARY MELANOTIC SARCOMA OF THE ESOPHAGUS

REPORT OF A CASE

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Primary malignant melanotic tumors of the esophagus are extremely rare, as evidenced by the fact that only three have been found in the literature, namely those of Baur (1), Joliat (2), and Voss (3). Metastatic melanomas however, are not unusual in the gastro-intestinal tract, where they may involve either the mucosa or submucosa.

In the present case, as well as in those reported previously, the diagnosis of esophageal tumor was not made until after death, since there were no signs of esophageal obstruction.

CASE REPORT

J. R., a man aged sixty-nine, entered St. Luke's Hospital July 6, 1933, complaining of a feeling of fullness in the epigastrium and the expectoration of large amounts of mucus, over a period of six weeks. Expectoration was usually accompanied by some coughing, and occurred a few minutes after eating. There had been a loss of forty pounds in weight during this time.

Thirty years before admission the patient had punctured the left eyeball. This healed promptly and produced a scar of the right cornea, which caused complete blindness in that eye. Since the healing of the eye there had been no further change in its appearance. Other than this the patient had always been in good health.

Careful examination revealed no cutaneous scars or nevi. There was a healed gray scar involving the entire left cornea. Except for a moderate amount of emphysema, the heart and lungs were normal. The spinous process of the fourth lumbar vertebra was quite prominent. Both ankle jerks and the right knee jerk were absent. The blood pressure was 120/70 and the Wassermann reaction was negative. The blood count, blood chemistry, urine and stool examinations all showed essentially normal findings.

An admission roentgenogram of the chest was normal, but one of the lumbar spine showed advanced osteoarthritic overgrowth, and the fourth lumbar vertebra slipped posteriorly. There was destruction of the anterior aspect of the body of the fifth lumbar vertebra. These findings were interpreted as being due to a metastatic tumor, and an unsuccessful attempt was made to find the primary focus. A Levine tube was passed into the stomach with ease, and the alcohol test meal showed no free hydrochloric acid and a total acidity of only 7. Two series of x-rays of the gastro-intestinal tract, following the administration of an opaque medium, were negative. Two attempts at examination of the large bowel were unsuccessful due to the patient's inability to retain the medium. Examination of the urinary tract following the intravenous administration of an opaque medium showed evidence of function on both sides.

Further x-ray examination revealed destruction in the fourth cervical vertebra, and twenty-two days after admission a roentgenogram of the chest gave evidence of metastases in the lower half of the right lung. One month after admission there was x-ray evidence of fluid in the right chest and 700 c.c. of this fluid were removed. It was not bloody; the cell count was 55 per cubic millimeter, with 4 per cent polymorphonuclear leukocytes and 96 per cent large lymphocytes. The specific gravity was 1.012 and the albumin (Esbach) 8 grams per liter. Proctoscopic and laryngoscopic examination revealed normal structures.
Symptomatic treatment was resorted to in an effort to keep the patient comfortable, and he was given five deep x-ray treatments over the spine. The course, however, was steadily downhill and death occurred on Aug. 16, 1933, forty-one days after admission.

An autopsy was performed ten hours after death. The anatomic diagnosis was primary melanotic sarcoma of the esophagus with metastases to the lungs, liver, pancreas, spine and regional lymph nodes.

On the posterior wall of the esophagus, 6 cm. above the cardia (Fig. 1), was a tumor mass 10 cm. in length, 4 cm. in width, and raised about 1.5 cm. above the surface. Surrounding the large tumor were many small solitary masses, the largest of which measured 2 cm. in diameter and was 4.5 cm. above the upper margin of the main tumor. The circumference of the esophagus directly above the tumor measured 4.5 cm. and just below it 5.5 cm. There was no evidence of a diverticulum.

The surface of the tumor was smooth and had a gray appearance. On section it was firm, rather friable, and uniformly gray in color. Directly behind the tumor were many large, grayish black mediastinal nodes which were firm on section.

The pleural surfaces of both lungs were studded with small, firm, grayish pink tumor masses measuring from 0.1 to 0.5 cm. in diameter. Nodules of the same character were present throughout the parenchyma of both lungs.

The liver weighed 2150 grams and contained many small gray and white tumor masses, the largest of which measured 1.5 cm. in diameter. Some of the nodules were very black, while others were nearly white. All were soft and friable.

The pancreas weighed 100 grams, and the head contained many small nodules similar to those found in the liver.

The medullary portions of the bodies of the 2nd, 3rd, and 4th lumbar vertebrae were Sections of the esophageal tumor were very cellular. The surface epithelium was intact in most places. The tumor cells were arranged along very thin connective-tissue septa, which usually contained a few blood and lymph vessels (Fig. 2). The cells were often piled up three or four deep; they were usually polygonal but varied in size and shape, and the nuclei were generally round and contained an abundance of chromatin material. Many mitotic figures were present in all the fields. With hematoxylin and eosin the majority of the cells showed no pigmentation, but along the septa and at the periphery of the necrotic areas were many cells filled with coarse black granules. However, with Bielschowsky's silver stain the nuclei of all the tumor cells stained a deep black, and scattered throughout the cytoplasm were many very fine black granules. The coarse pigment granules in the large cells also took a deep black stain. Iron stains showed the uniform absence of this metal in all the tumor cells; it was present, however, in small quantities along the vessels of the septa. Pieces of the melanotic tissue placed
in 10 per cent and also in concentrated hydrochloric, sulphuric, nitric, and acetic acids showed no loss of color after treatment for twenty-four hours. The same can be said for the treatment of the tissue with alkalies: potassium and ammonium hydroxide. Rapid fading of the color of the pigment took place, however, when thin pieces of the tumor were placed in hydrogen peroxide. The pigment did not stain with any of the fat stains. The cellular morphology was essentially the same in the metastases as in the primary tumor. However, the cells in the liver metastases stained more deeply than the cells in the primary tumor.

FIG. 2. PRIMARY MELANOTIC SARCOMA OF ESOPHAGUS

DISCUSSION

In none of the reported cases of melanic sarcoma of the esophagus has esophageal obstruction been a prominent feature. In the cases of Voss (3) and Baur (1) there was complaint of pain on swallowing, but there was never any vomiting. The tumors all occurred in elderly individuals; two of these were males; the sex of the other was not reported. There were widespread metastases in all cases except that of Baur.

The origin of the pigment melanin as well as the cells in which it resides is still a matter of controversy. The histological evidence of Unna (4), Judalewitsch (5) and dalla Favera (6) for the origin of pigmented nevus cells from the rete mucosum of the epithelium is equally good as the theoretical evidence produced by Ribbert (7) for their origin from mesothelial chromatophores. Masson (8), from his careful researches, concludes that the epidermoid melanoblasts resemble very closely the tactile cells and neuroglia of the end apparatus of the sen-
sory nerve filaments of the skin. Kromayer (9) believes the nevus cells have an epidermoid origin, but his work indicates that they undergo metaplasia into mesothelial cells.

Melanotic tumors arising in the gastro-intestinal tract may have their origin from the pigmented cells which are found abundantly in the submucosa. Tuczek (10) and Voss (3) believe they arise from the ectodermal sympathetic nervous components found in the submucosa. Lubarsch (11), on the other hand, feels that all melanotic tumors of the gastro-intestinal tract are really metastatic growths from a nevus elsewhere in the body.

Neither the origin nor the chemical structure of melanin has been settled, and in all cases it is necessary to distinguish melanin from lipofuchsin and hemosiderin. With the usual hemotoxylin and eosin stain this cannot be done. With Bielschowsky's silver stain melanin stains a deep black, and is thus distinguished from other pigments, which remain unstained. Melanin differs from hemosiderin in that it does not give a positive iron reaction, but in necrotic portions of a tumor where there has been hemorrhage the tumor cells may take up some of the iron-bearing pigments.

Melanin is stable to both dilute and concentrated mineral acids and alkalies. It is susceptible to the oxidizing agents such as hydrogen peroxide and permanganate. The pigment is not stained by the usual fat stains, but it does blacken when treated with osmic acid.

Melanin probably arises by virtue of the action of an enzyme or ferment on some colorless precursor which is presumably a protein or portion of a protein molecule. It seems certain that it has a metabolic rather than a hematogenous origin.

**Summary**

A case of primary melanotic sarcoma of the esophagus, undiagnosed before autopsy, is described in a man of sixty-nine. The post-mortem examination showed a primary pigmented tumor of the esophagus with metastases to lungs, liver, pancreas, and spine. The presence of melanin in the tumor cells was proved by the use of silver, iron, and fat stains, as well as by treatment of the tumor tissue with hydrogen peroxide, acids, and alkalies.

**Bibliography**